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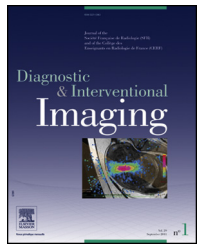

E-QUID: ANSWER /*Gastrointestinal imaging*

Epidermoid splenic cyst☆

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Case study

A 22-year-old woman from North Africa was referred to the Emergency Department. She complained from left-sided abdominal pain that appeared three days before. She had no remarkable medical history except a full-term pregnancy with vaginal birth nine months before and she had not recently travelled.

Clinical examination revealed a soft abdomen with a localized tenderness in the left upper quadrant and perceptible splenomegaly. The patient had no fever. The initial laboratory test results were normal apart from thrombocytopenia (88,000 platelets/mm^3). Biological tests showed no inflammation and results of liver function tests and lipase tests were normal. Urinalysis was negative.

Abdominal ultrasonography examination (Fig. 1) was first performed followed by a computed tomography (CT) examination of the abdomen and pelvis before and after intravenous administration of iodinated contrast material (Fig. 2).

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☆ Here is the answer to the case An uncommon cause of left upper abdominal pain previously published. As a reminder we publish again the entire case with the response following.
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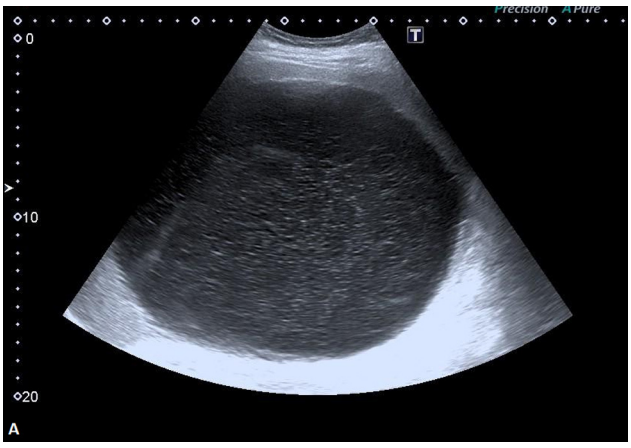

Figure 1. Ultrasonography examination of the abdomen.

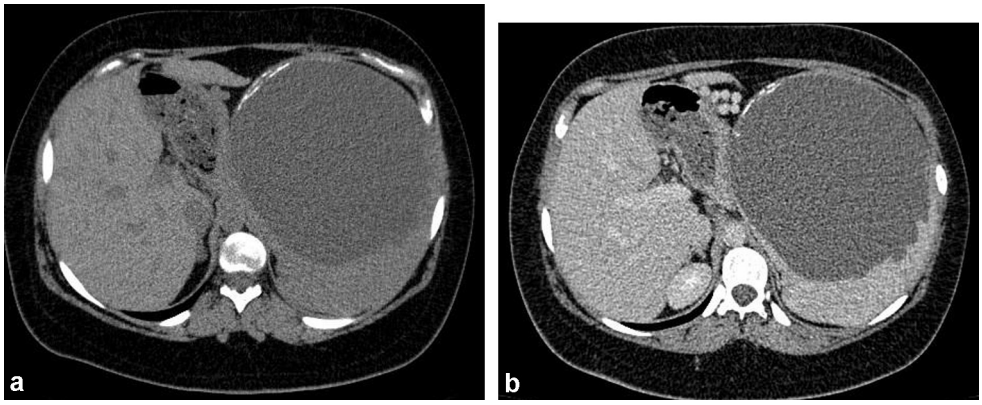

Figure 2. CT examination of the abdomen in the axial plane, (A) before and (B) after intravenous administration of iodinated contrast material during the enteric phase.

What is your diagnosis?

On the basis of clinical and imaging findings, which diagnosis is the most plausible from among the following:

- splenic hydatid cyst;
- splenic epidermoid cyst;
- splenic pseudocyst;
- splenic cystic lymphangioma.

Diagnosis

Epidermoid splenic cyst.

Comments

Abdominal ultrasonography (Fig. 3) demonstrated a well-circumscribed large mass with thin walls, slightly echogenic content and posterior acoustic enhancement, located in the left upper quadrant.

Abdominal CT examination (Fig. 4) demonstrated a homogenous fluid-containing splenic lesion with wall calcifications (solid arrow) and scalloped edges at its posterior aspect (dotted arrow). There were no septations, membranes or vesicles within the cyst. The splenic vein was compressed, although it remained patent, leading to segmental portal hypertension (arrow head). The liver and pancreas were normal.

A repeat and more targeted ultrasonography examination was carried out the following day (Fig. 5), which demonstrated mobile internal echoes and confirmed the presence of peripheral trabeculations (solid arrows), a characteristic feature of splenic epidermoid cyst.

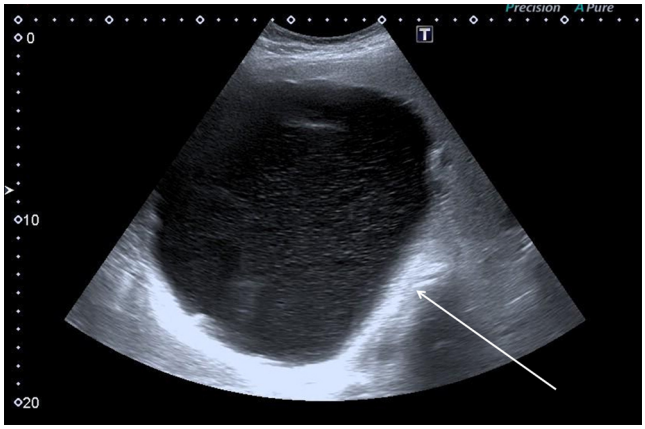

Figure 3. Abdominal ultrasonography shows amass with multiple internal echoes and posterior acoustic enhancement.

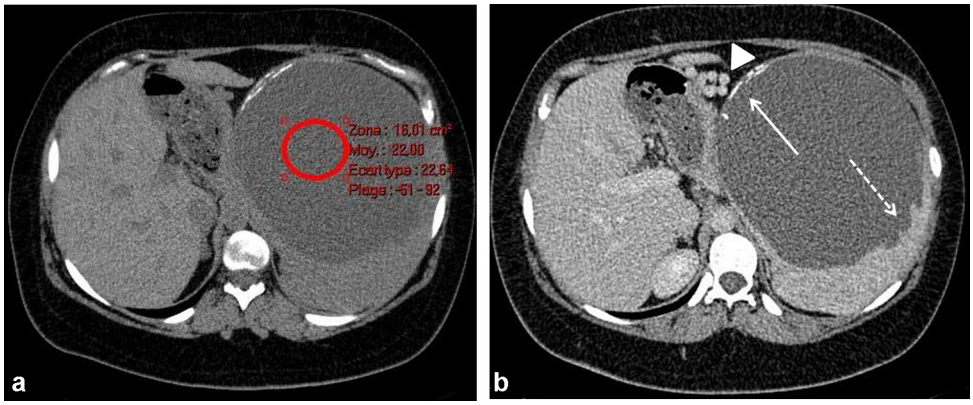

Figure 4. CT examination of the abdomen in the axial planes. a: splenic mass with fluid content (22 HU); b: wall calcifications (solid arrow) and peripheral scalloping (dotted arrow). Segmental portal hypertension due to splenic vein compression (arrow head) is visible.

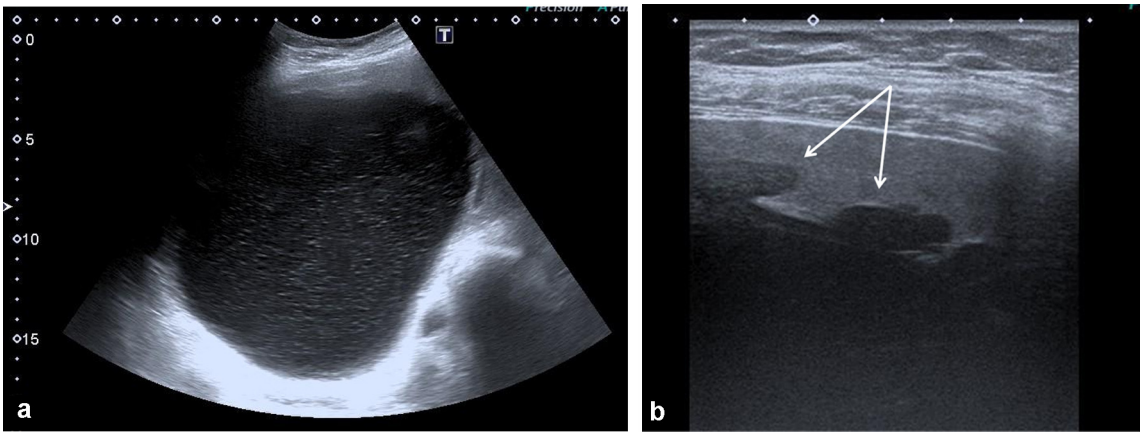

Figure 5. Targeted ultrasonographic examination of the splenic mass shows: (a) mobile internal echoes and (b) scalloped margins (solid arrows).

The clinical presentation in association with the results of laboratory test (no recent travels and negative hydatid serology) together with the imaging features (no septation, presence of vesicles or membrane within the cyst and normal appearance of the pancreas) allowed the definite diagnosis. The patient underwent surgical cystectomy through laparoscopy. On gross examination, the cyst had an internal surface made up of whitish trabeculations peripherally that formed a mesh (Fig. 6) and clear, greenish intracystic fluid. The diagnosis of epidermoid splenic cyst was further confirmed histologically.

Discussion

Splenic epidermoid cysts are benign congenital tumours of the spleen, defined histologically by the presence of a stratified epithelial lining that may be either squamous or

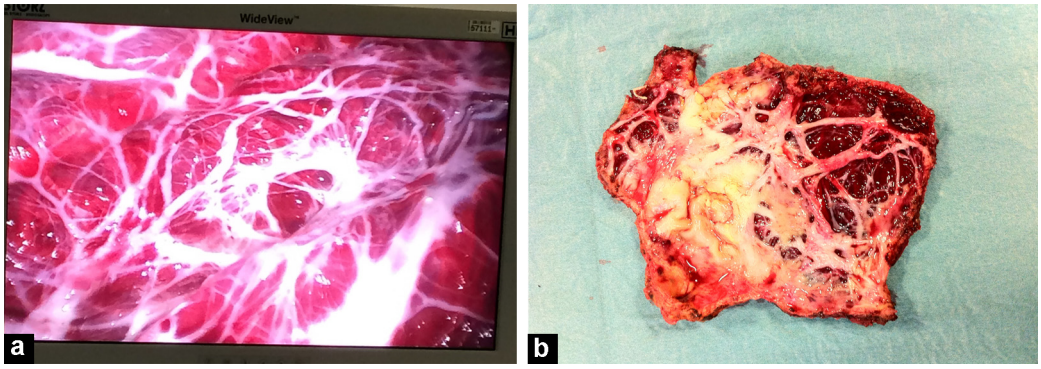

Figure 6. Photograph shows macroscopic view of the cyst. a: during laparoscopy, the cyst presents with white streaks on its internal surface; b: resected specimen.

cuboidal, and they account for between 10 and 25% of non-parasitic splenic cysts and 2.5% of all cystic lesions of the spleen [1]. They are most commonly seen in children and young adults, with a predilection in females. The majority are discovered incidentally because they are asymptomatic, though they can become symptomatic if they are large or complicated by infection, rupture or haemorrhage [2]. As a rule, they present as single lesions whereas multiple forms are very rare, and they can vary in size from 1 to over 25 cm in diameter. They are usually located at the upper pole of the spleen and are typically subcapsular. At gross examination, the internal surface of the cyst wall is smooth and whitish with numerous peripheral trabeculations. They contain clear, and sometimes thick, fluid [3].

Ultrasonography is the most useful examination to reach the correct diagnosis. An epidermoid cyst has a liquid content with posterior acoustic enhancement and characteristically contains multiple internal echoes that are typically mobile, and are attributed to the presence of cholesterol crystals and keratin particles. In case of intracystic haemorrhage and/or secondary infection, a fluid-fluid level may be seen. The margins are typically scalloped in places due to the wall trabeculations. The adjacent splenic tissue is sometimes enlarged due to venous compression.

On CT, the content of epidermoid cysts is of varying density, sometimes fatty (between −80 and −130 HU), and sometimes fluid (between 10 and 30 HU) or showing higher HU if there is intracystic haemorrhage. Non-specific peripheral calcifications are often present.

MR imaging can be performed when ultrasonographic and CT findings are doubtful, or for malignant and/or atypical lesions. Epidermoid cyst shows features that are common to all cystic lesions, with low signal intensity on T1-weighted sequences and marked high signal intensity on T2-weighted sequences indicating fluid density, and no enhancement after gadolinium-chelate administration. High signal intensity on T1-weighted MR images indicates intracystic haemorrhage [4].

Symptomatic cysts require surgery, and an attempt should be made to take the most conservative approach (cystectomy or partial splenectomy) in young patients in whom the aetiology is certain and/or all potentially malignant or dangerous pathology (hydatidosis) has been excluded. Percutaneous fluid drainage prior to or during surgery in epidermoid cyst assists in maintaining a conservative approach but is associated with a higher rate of early recurrence [2,5].

The differential diagnoses include:

- hydatid cyst: in our patient, there was no indication of contagion, and hydatidosis serology was negative although this is not always informative because it is positive in only 80% of cases. In the early stage (Gharbi I), the wall is thick and made up of three layers although it can still be irregular and calcified. On imaging, the content is more heterogeneous with, in later stages, hydatid daughter cysts or detached secondary intracystic membranes, a pathognomonic imaging sign for hydatid cyst [6]. This diagnosis is a formal contraindication for fenestration;
- intrasplenic pseudocyst in the peripartum period: some authors believe this to be related to a fragile spleen during menstruation and pregnancy, most often secondary to a trauma, a local inflammation (following pancreatitis or tuberculosis) or a splenic infarction. In this case, there was no indication of trauma to the spleen or history of pancreatitis. On imaging, the content usually tends towards anechoic although there can be low-level internal echoes and wall calcifications;
- cystic lymphangioma: this condition is less common and rarely unilocular, except when it is small. In our patient, imaging did not show any satellite cystic formation.

Conclusion

The diagnostic approach of splenic cysts must take into account the clinical context and imaging features. An accurate preoperative diagnosis helps avoid unnecessary splenectomy or even a risky fenestration. Ultrasonography allows the diagnosis of epidermoid cyst by demonstrating mobile internal echoes.

Disclosure of interest

The authors declare that they have no conflicts of interest concerning this article.

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